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(FILE 'HOME' ENTERED AT 13:22:30 ON 24 JUL 2005)

FILE 'CAPLUS, MEDLINE, BIOSIS' ENTERED AT 13:22:42 ON 24 JUL 2005

L1 985 S HUNTINGTONS DISEASE
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L4 6 S L1 (P) PREVENT?
L5 20 S L2 (P) PREVENT?
L6 0 S L1 (P) VACCINE
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FILE 'STNGUIDE' ENTERED AT 13:32:42 ON 24 JUL 2005

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Diseases and Conditions

Huntington's disease

From MayoClinic.com
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Overview

Huntington's disease (Huntington's chorea) is a progressive, degenerative disease that causes certain nerve cells in your brain to waste away. As a result, you may experience uncontrolled movements, emotional disturbances and mental deterioration. The disorder was documented in 1872 by American physician George Huntington. The name "chorea" comes from the Greek word for "dance" and refers to the incessant quick, jerky, involuntary movements that are characteristic of this condition.

Huntington's disease is an inherited disease. Signs and symptoms usually develop in middle age, and men and women are equally likely to develop the condition. Younger people with Huntington's disease often have a more severe case, and their symptoms may progress more quickly. Rarely, children may develop this condition.

An estimated one in 10,000 Americans has Huntington's disease, with about 30,000 known cases in the United States. About 150,000 Americans may be at risk of inheriting Huntington's disease from a parent.

Medications are available to help manage the signs and symptoms of Huntington's disease, but treatments can't prevent the physical and mental decline associated with this condition.

Signs and symptoms

The earliest signs and symptoms of Huntington's disease often include personality changes and decreased cognitive abilities. You may demonstrate symptoms such as irritability, anger or paranoia or

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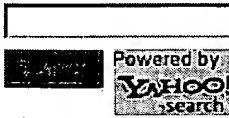
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show signs of depression. You may also begin to have difficulty making decisions, learning new information, answering questions and remembering important information. Your family and friends may notice these changes before you become aware of them.

- Cerebral palsy
- Essential tremor
- Huntington's disease
- Benign paroxysmal positional vertigo (BPPV)
- Tourette syndrome

Early physical signs and symptoms of Huntington's disease may include mild balance problems, clumsiness and involuntary facial movements such as grimacing. As the disease progresses, you may

- Sudden jerky, involuntary movements (chorea) throughout your body
- A wide, prancing gait
- Severe problems with balance and coordination
- Difficulty shifting your gaze without moving your head
- Hesitant, halting or slurred speech
- Inability to swallow
- Dementia

Young people who develop Huntington's disease may have symptoms that mimic Parkinson's disease: muscle rigidity, tremors and slow movements. Those with early-onset Huntington's disease also may

The disease usually develops slowly, and the severity of signs and symptoms is related to the degree of gene abnormality. Death occurs about 10 to 30 years after signs and symptoms first appear.

- Dementia: Not always Alzheimer's
- Brain & Nervous System

Causes

Huntington's disease is an inherited condition caused by a single abnormal gene. Doctors refer to the autosomal dominant disorder because only one copy of the defective gene, inherited from either parent, can produce the disease. If one parent has the single faulty gene, the chance that an offspring will have the disease is 50 percent. Because signs and symptoms typically appear in middle age, some parents may not know they have the gene until they've already had children and possibly passed on the trait.

If your child doesn't inherit the faulty gene, he or she won't develop Huntington's disease and can't pass it on to the next generation. Everyone who has the gene eventually develops Huntington's disease, if he or she lives long enough.

Risk factors

If one of your parents has Huntington's disease, you have a 50 percent chance of developing the disease. In some cases, you may develop Huntington's disease without having a family history of the condition. Such a case may be the result of a genetic mutation that happened during your father's sperm development.

- Compiling your family medical history: Use the past to prepare for future

When to seek medical advice

See your doctor if you notice changes in your movements, emotional control or mental ability. These symptoms can be the result of many conditions, so they don't necessarily mean you have Huntington's disease.

If you have a family history of Huntington's disease, you may want to talk with your doctor. Some people undergo genetic testing to see whether they carry the defective gene. That can help determine whether you or potentially your children — are at risk.

Deciding whether to be tested for the gene is a personal decision. For some people, the uncertainty of carrying the faulty gene is stressful and distracting. For others, the knowledge that they will develop the disease is burdensome. If you're uncertain whether testing is right for you, consider contacting a genetic counselor who specializes in medical genetics can help you understand the implications of a positive or negative test result, guide you through the testing process, and help you weigh the pros and cons. Ask your doctor for help locating a counselor.

If you choose to be tested, consider paying for it with your own money so that the test results remain confidential.

- Genetic testing: Weighing its benefits and risks

Screening and diagnosis

To determine whether you may have Huntington's disease, your doctor performs a physical exam and a medical history and that of your family. He or she may also ask about any recent emotional or intellectual changes you have had. A computerized tomography (CT) or magnetic resonance imaging (MRI) scan may show a cross-section of your brain's structure. Your doctor may suggest a blood test to determine whether you carry the defective gene.

- Video: Computerized tomography (CT) scan
- Computerized tomography
- MRI: Viewing the body's hidden structure

Complications

After onset of the disease, signs and symptoms continue until death. Though the signs and symptoms vary from person to person, vital functions such as swallowing, eating, speaking and walking usually degenerate over time. Some people with Huntington's disease develop depression, and some are at risk of suicide. However, death is usually the result of complications of the disease, such as an infection or a fall.

- Depression
- Understanding suicide: Know the signs

Treatment

Most people who have Huntington's disease eventually become physically and mentally disabled. As the disease progresses, long-term nursing home care may be necessary.

No satisfactory treatment is available to stop or reverse Huntington's disease, but some approaches can help control the symptoms.

Medications

Tranquilizers such as clonazepam (Klonopin) and antipsychotic drugs such as haloperidol (Haldol) or risperidone (Risperdal) can help control movements, violent outbursts and hallucinations.

Various medications, including fluoxetine (Prozac, Sarafem), sertraline (Zoloft) and nortriptyline (Aventyl), can help control depression and the obsessive-compulsive rituals that some people with Huntington's disease experience. Medications such as lithium (Eskalith, Lithobid) can help control extreme emotions and mood swings.

Side effects from many of the drugs used to treat the symptoms of Huntington's disease may include fatigue and restlessness. In some instances, antipsychotic drugs may cause side effects that mimic those of Parkinson's disease, including involuntary twitching in your face and body (tardive dyskinesia).

Speech therapy

Huntington's disease can impair your speech, affecting your ability to express complex thoughts. You can use speech therapy to help. Remind friends, family members and caregivers that if you don't speak, it doesn't mean that you don't understand what's going on. Ask people to continue talking to you and keep your speech as normal as possible.

Prevention

If you have a family history of Huntington's disease, you may want to consider genetic counseling before having a family. A blood test can determine the presence of the faulty gene, even before you show signs or symptoms. If a parent carries the defective gene, his or her child has a 50 percent chance of developing Huntington's disease.

If you're at risk of passing the genetic defect that causes Huntington's disease to your children, you may want to consider adoption or certain forms of assisted reproduction. One possibility is in vitro fertilization with preimplantation genetic screening. In this procedure, embryos are screened for the Huntington's disease gene mutation, and only those without the mutation are then implanted in the woman's uterus.

Self-care

Having Huntington's disease presents a number of challenges that require daily or regular attention. The following steps may improve how you feel:

- **Exercise regularly.** People with Huntington's disease who exercise regularly tend to fare better than those who don't exercise, and physical activity can help you feel better mentally and physically. Keep your hands away from sharp, hard objects in case you fall while moving around. Consider wearing special padding to protect your head from falls that may happen while you're on walks. Wearing sturdy, properly fitting shoes may help prevent falls.
- **Maintain proper nutrition.** People with Huntington's disease may burn as many as 5,000 calories a day. Be sure to get adequate nutrition to maintain your body weight, and consider extra vitamins and minerals. Because Huntington's disease can impair coordination, you may need assistance when eating. Take time for meals. Cutting food into small pieces or eating pureed food may make swallowing easier. Avoid choking. Dairy products may make you secrete excess mucus, which may increase your risk of choking. Occupational therapists may make other suggestions on how to improve your swallowing. Use adaptive devices and suction cups and tableware designed for people with disabilities may prevent spills.
- **Drink plenty of fluids.** Huntington's disease can make you vulnerable to dehydration. Drink plenty of fluids, especially during hot weather. Some people find that bendable straws make drinking easier.

- Water: How much should you drink every day?

Coping skills

Most people with Huntington's disease eventually need assistance in performing daily functions. You may be able to live on your own but can't take care of tasks necessary for independent living. In these cases, a group-home or assisted living facility can provide a safe and comfortable living situation that allows you to maintain your independence.

Tips for the caregiver

If you're a family member or friend of someone with Huntington's disease, caregiving can be time-consuming and exhausting. But certain resources can make caregiving easier. Hiring someone to help with household tasks or physically caring for the person with Huntington's disease is one option. In addition, many local, state and federal agencies provide such assistance as meal programs, occupational therapy, nursing assistance and counseling.

If you're a caregiver, take time to relax and take care of your own health. Therapeutic recreation and support groups can provide a place for someone with Huntington's disease to interact with others and pursue interests and hobbies. Respite care also provides home caregivers a break.

Some cases of Huntington's disease may progress to the point where friends and family members cannot provide adequate care. Long term care facilities can provide a safe and comfortable living situation when home care isn't a viable option.

May 11, 2005

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